



ACC.15

TCT@ACC-12 | innovation in intervention

A640
JACC March 17, 2015
Volume 65, Issue 10S

FIT Clinical Decision Making

UNUSUAL RIB NOTCHING IN TETRALOGY OF FALLOT

Poster Contributions

Poster Hall B1

Saturday, March 14, 2015, 3:45 p.m.-4:30 p.m.

Session Title: FIT Clinical Decision Making: Structural Heart Disease and Pulmonary Hypertension

Abstract Category: Congenital Heart Disease

Presentation Number: 1142-146

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Background: Tetralogy of Fallot (TOF) consists of subpulmonic stenosis (PS), ventricular septal defect (VSD), overriding aorta and right ventricular hypertrophy (RVH). Rib notching is not a feature of TOF and can be due to other vascular abnormalities.

Case: A 39-year-old man with an unrepaired TOF presented with exertional dyspnea and cyanotic spells. Examination revealed lip cyanosis and clubbing of fingers. Cardiac auscultation noted a grade 4 systolic murmur. Electrocardiogram showed right axis deviation and RVH with strain. Chest radiograph noted a boot-shape cardiac silhouette, a lacy reticular vascularity in bilateral lower lungs and notching of fourth and fifth right posterior ribs.

Decision Making: Rib notching is not a feature of TOF and can be due to other vascular abnormalities. Echocardiograms noted RVH, PS, VSD and an overriding aorta consistent with TOF. Cardiac MRI confirmed the findings of TOF. Magnetic resonance angiogram (MRA) noted severe PS and left pulmonary arterial atresia. There were substantial major aortopulmonary collateral arteries (MAPCAs) with an intrapulmonary course perfusing left lung and right lower lungs (caused lacy reticular vascularity in chest radiograph). Right subclavian artery was stenotic with collateral arteries arising proximally with an extrapulmonary intercostal course to perfuse right lung and caused an unilateral rib notching. These well-developed collateral circulation alleviated his symptoms for decades. He declined surgical intervention and was followed-up with echocardiographic examination every year.

Conclusion: Rib notching in TOF is unusual and cardiac MRI is important in defining the congenital defects as well as the intrapulmonary and extrapulmonary collateral circulation in this case.